Labio-Glosso-Laryngeal Paralysis.

Wm. W——, aged 58 years, began to complain in May, 1869, that the saliva dribbled from his mouth when speaking or reading. One month afterward he said he was conscious of a difficulty in the pronunciation of words, although at this time it was so slight as not to be perceptible to those with whom he conversed. The following autumn there was a very evident hesitation in his speech; he talked slowly and deliberately, but correctly, excepting a defect in the articulation of certain words, and his voice had assumed a nasal tone.

In February, 1870, he visited some friends in the northern part of New York, and was able to travel alone. While in the country he placed himself under the charge of a physician, and electricity was employed but without benefit, as in his letters to his family he stated that he was growing worse.

He returned to Albany, in June, 1870, in the following condition: complete loss of speech; difficulty in deglutition, any effort to swallow being attended with paroxysms of coughing and suffocation; paralysis of the tongue, which could only be protruded a quarter of an inch beyond the edge of the teeth; body well nourished and appetite good; partial loss of motion in both arms, especially on the left side; pain in the thumbs, arms, and back of the neck; mental action unimpaired, except that the emotions were easily excited so that he laughed and wept without sufficient cause; could write perfectly with his elbow resting upon the table, but was unable to raise his arm so as to feed himself; could walk without difficulty.

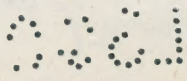
In July it was noticed that he dragged his feet a little, but he could still walk alone for a considerable distance. After this time, the movements of the arms and legs gradually became weaker, until in November he was unable to get up and down stairs, while at the same time the difficulty in deglutition increased. His writing became more and more illegible, and in the latter part of November he had to give it up altogether, and communicated his wishes to others by pointing out letters with a stick placed in his hand for the purpose.

In December he had a severe chill, which was followed by acute pain and complete loss of motion in the left arm, which became œdematous, and remained so until a few days before his death. He could walk only a few steps, and had great difficulty in doing even this.

During December the flexor muscles of his hands and arms began to contract, and in the latter part of the month his feet became œdematous. Sight, hearing, and appetite remained good throughout.

I saw the patient through the politeness of Dr. Fowler, his attending physician, from whom I obtained the above notes, on the morning of January 4th, 1871.

He was then sitting in a chair propped up by pillows, being unable to lie down on account of dyspnoea; complete loss of motion, except a slight nodding of the head and a little movement of the right hand; sight and hearing appeared unimpaired; speech entirely lost; mouth partly open and lips immoveable, except a slight twitching of the left angle of the mouth; cheeks flaccid; tongue completely paralyzed and lying on the floor of the mouth; respiration feeble and occasional coughing; pulse 90 per minute and regular; both arms paralyzed and slightly flexed and attempts to straighten the fingers caused pain; lower extremities completely paralyzed, and feet and ankles œdematous; defecation



natural; micturition slow and frequent; attempts to swallow occasioned distressing cough and suffocation, and the aliments were often rejected through the nose.

He remained in this condition until the afternoon of the same day, when an attempt to swallow some porridge brought on severe coughing and strangling. At seven o'clock that evening he died without a struggle.

Autopsy twenty hours after death.

External appearances. Rigor mortis well marked. Body spare, but not emaciated. No very evident signs of muscular atrophy.

Head. Scalp very dry. Scull cap removed with great difficulty, owing to adhesions of the dura-mater, which was torn in trying to separate it from the bone. Dura-mater very much thickened. Arachnoid normal, with considerable serous infiltration of the sub-arachnoidean connective tissue. Pia-mater much injected. The cerebral substance both cortical and medullary appeared to be of normal color and consistency, but exceedingly hyperæmic. The following condition of the cranial nerves was found. 1st. Olfactory, normal. 2d. Optic, normal. 3d. Motor oculi, normal. 4th. Patheticus, small. 5th. Trigeminus, on the left side flattened, gray and softened; on the right side, larger and very hyperæmic. 6th. Abducens atrophied, especially on the left side. 7th. Facial atrophied, and gray on both sides. 8th. Auditory, normal. 9th. Glosso-pharyngeal, normal. 10th. Pneumogastric, atrophied on both sides. 11th. Spinal accessory much atrophied. 12th. Hypoglossal so much atrophied on both sides as to resemble mere threads or filaments of connective tissue. The corpora striata and optic thalami were normal. The cerebellum was very hyperæmic, but otherwise presented nothing unusual. The pons Varolii and medulla oblongata appeared to be of firmer consistency than usual.

Spinal cord. Spinal meninges much injected. The anterior spinal roots were atrophied especially on the left side. Transverse sections of the cord showed the anterior cornua of gray matter, as well as the left anterior and right lateral columns, to be of a dark rose color as if very hyperæmic.

Thorax and abdomen were not examined on account of objections on the part of relatives of the deceased.

Portions of the brain, cerebellum, and spinal cord were immersed in absolute alcohol, preparatory to making sections for microscopic examination. When sufficiently hardened thin sections were made, stained with carmine, rendered transparent with benzole, and mounted in balsam.

The sections of the brain revealed nothing abnormal. The sections of the cerebellum a very hyperæmic condition of the part and a granular degeneration of the large ganglionic cells forming the middle layer of the cortical portion.

Thirty sections were made at various levels of the medulla oblongata involving the roots and nuclei of implantation of the cranial nerves, especially those of the facial and hypoglossal. Careful microscopic examination of these specimens with objectives varying from 15 to 900 diameters, demonstrated that the portion of medulla forming the floor of the fourth ventricle was the seat of several pathological lesions.

There was a decided hypertrophy or overgrowth of the connective tissue, which appeared to have encroached upon and to some extent replaced the several groups of ganglionic cells which form the nuclei of implantation for the facial and hypoglossal nerves. The individual cells comprising these groups were separated from one another and in many instances had lost their stellate appearance, their radiating processes

having been destroyed so that each cell remained isolated and disconnected from its neighbors. These cells had also undergone a degenerative process which in many cases rendered them simply a collection of fine granules, and a deposit of brownish yellow pigment had taken place to such an extent as to give the cells an appearance almost precisely similar to those which are normally found in the locus niger of Soemmering; they were fewer in number than usual and diminutive in size.

Sections of the cord made in the cervical, dorsal, and lumbar regions showed a sclerosis with increase of connective tissue in the anterior and lateral columns, which was most marked in the left anterior and right lateral columns. The multipolar ganglionic cells situated in the anterior cornua of gray matter were fewer in number than usual, and many of them appeared granular and very much pigmented.

The form of disease of which the above is a typical example, has been described by Trousseau and Duchenne, and a number of cases have been reported in various medical journals. It commences with a paralysis of the lips and tongue which is followed by a paralysis of the soft palate and pharynx; in many cases the disease does not stop here, but gradually attacks the muscles of the trunk and extremities. There is no muscular atrophy, but simply a loss of motor power. In those cases where an autopsy has been made, the roots of the hypoglossal and some of the other cranial nerves have been found to be atrophied, but little or nothing has been said of the condition of the central origin of these nerves, except in one case described by Dr. Samuel Wilkes, in the last volume of Guy's *Hospital Reports*. Dr. Wilkes says that "there was very obvious gray red change of the calamus scriptorius

from the nib upward and outward for half an inch." This change would necessarily implicate the cells of implantation of the facial and hypoglossal and probably of the spinal accessory roots, and we therefore have a central lesion accounting for the peripheral paralysis. Dr. Wilkes also states that the anterior half of the white matter of the cord was hardened and atrophied. In the case above reported not only the anterior, but also the lateral columns of the cord were affected so that we have a descending degeneration of the motor tracts of the cord, consecutive to a primary lesion situated in the medulla. This is fully in accordance with the views presented by Bouchard in his work on secondary degenerations of the spinal cord, and accounts for the progressive paralysis of the trunk and extremities which follows the original loss of motion in the lips, tongue, and palate.

An interesting fact in connection with this secondary disease of the cord, is the ultimate permanent contraction of the flexor muscles. This contraction with rigidity was present in the case reported, and is also mentioned by Trousseau as having occurred in one of his cases. Bouchard speaks of it as an almost constant symptom of secondary sclerosis of the anterior columns of the cord.

This case affords an illustration of the aid which the study of physiological anatomy may derive from pathology. We have here a very complete destruction of the cells of implantation of the facial nerves, and yet only a part of the muscles supplied by the facial are paralyzed. The orbicular muscles of the eye sare unaffected, and the patient can wink voluntarily while the buccinators and orbicularis oris are perfectly paralyzed. This can only be explained by the fact that some of the fibres of the facial nerve (as has been stated by

Stilling and Van Der Kolk) do not pass into their proper nucleus, but either pursue an upward course along the raphe of the cord, or else run into small ganglionic groups at a considerable distance from the nucleus proper. Thus from the fact of the non-paralyzed condition of the orbicularis oculi, notwithstanding the destruction of the nucleus of the facial nerve, there can be but little doubt that the fibres referred to by Stilling and Van Der Kolk are those which supply this muscle, and it is probable that these fibres terminate in ganglionic cells in the neighborhood of those from which the fifth pair of nerves originate, and thus we have an anatomical arrangement which may aid in the explanation of the reflex phenomenon of winking.

The prognosis in this disease is decidedly unfavorable. Among the many cases reported, not one case of perfect recovery is mentioned, and it is probable that the improvement which seems to have taken place in some of them was only temporary. The manner of commencement may be very sudden or slow and insidious, but there is but little variation in the ultimate termination. Death takes place either as a result of suffocation from the impaction of a bolus of food in the glottis, or else the slow accumulation of mucus in the lungs, which the patient can no longer cough up, gradually results in asphyxia. As for treatment there is little to be said. Life may be prolonged by the application of galvanism to the upper part of the cord and along the tracts of the affected nerves, but nothing as yet known has any power to retard the progress of the disease in the nerve centres. The following is a list of authors with references to where the cases are reported:

Trousseau—*Clin. Med.*, t. II., p. 334.

Duchenne—*Electrisation localisée Doc.*, p. 640; *Gaz. Hebd.*, VIII., 12, 1861.

Dumenil—*Gaz. Hebd. de Med. et de Chir.*, June, 1859, p. 390.

Samuelson—*Berl. Kl. Wochensch.*, No. 27, 1868 ; *Half-yearly Compend. of Med.*, January, 1869.

Ramskill—*Lancet*, June, 1869.

Herard—*Gaz. des Hopitaux*, No. 20, 1868 ; *Rankin's Abstract*, January, 1868.

Müller—*Hospit. Tidend*, Nos. 22 and 23, 1861 ; *Schmidt's Jahrb.*, vol. 113, p. 169 ; *Year Book of Medicine*, 1862.

Tommasi—*L'Union*, 114, 1862.

Wilks—*Guy's Hospital Reports*, vol. XV. ; *Rankin's Abstract*, July, 1870 ; *British Medical Journal*, March 19, 1870.